DERMOIDS OF THE MEDIASTINUM.

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The occurrence of mediastinal dermoids is infrequent, there having been but 72 cases reported in the literature. Rare as they are, however, when they are encountered the fate of the patient is largely dependent upon the surgeon's knowledge of their peculiarities. Inasmuch as such knowledge is dependent upon a familiarity of recorded cases, a tabulation of cases heretofore observed seems worth while.

The case reports collected from the literature, herewith appended, are many of them deficient in detail, yet they all contain some point of interest and are all included in this list, to which I am able to add one case of my own.

From these reports, one is able to formulate a fairly comprehensive life history of these tumors and to gain a fairly clear notion as to the symptom-complex that should suggest their presence. Though most of the tumors were first noted postmortem, a sufficient number have been subjected to operative treatment to furnish some idea of the line of procedure best adopted in the individual case.

My case is as follows: Mrs. W., aged twenty-three years; has always enjoyed good health. In November, 1914, she noticed some difficulty in respiration and had some sense of fulness in the neck. Soon afterward a bulging was noticed above the breast bone. She consulted several surgeons, who made a diagnosis of mediastinal sarcoma and refused treatment.

At examination a bulging in the suprasternal notch was apparent on inspection. The skin covering this area was slightly reddened. On palpation the tumor was slightly tender to touch and presented a soft semifluctuating resistance. The mass extended 2 cm. above the upper border of the sternum and was hidden by the sternomastoid muscles on either side. There was no bulging of the sternum or of the costal cartilages. On percussion there was dulness extending on either side of the sternal borders and downward as far as the angle.

Because of the reddened skin and boggy feel of that portion of the tumor accessible to palpation the diagnosis of dermoid was suggested, because of the close resemblance to the appearance and and consistency of irritated wens. This opinion was strengthened by the globular outline of the substernal dulness. Sarcoma was excluded because of the consistency of the palpable portion of the tumor and because only the upper portion of the mediastinal space was occupied by the tumor.

Operation was begun by exposing the upper portion of the mediastinum. A transverse incision was made over the upper border of the sternum, extending well beyond the insertion of the sternomastoid muscle on the left side. The insertion of the muscle was severed. The superior pole of the globular tumor was thus readily exposed. This was freely incised and a grayish-yellow greasy fluid escaped. After this was sponged out a mass the size of a walnut presented. This was covered with fine lanugo-like hair of the color of a newly hatched goslin. The appearance of this mass established the diagnosis without question.

The operation was completed by the removal of the mass and the exsection of as much as possible of the sac. The part adjacent to the sternum was readily removed; that of the lateral borders caused greater apprehension. The posterior wall of the cyst was in close apposition with the large vessels of the neck, and was allowed to remain. The cavity was swabbed out with iodin and the wound was closed except for a small opening admitting a drain. This was removed in a week and the wound rapidly became closed, and has remained so.

The mass removed presented a dermal surface studded with the fine hair above noted. On section the mass showed a fatty tissue, save for the epidermal covering. On microscopic examination stratified squamous epithelium and sweat and sudoriferous glands with hair follicles were noted.

The cyst wall was of the same structure, save that hair was much less abundant and the glands sparse. There were no more highly organized tissues present.

AGE. The majority of the cases have been observed in early adult life, the largest number being noted between the ages of twenty and thirty. The extremes of life are not exempt, one case having occurred in infancy, and another at four and one-half years, and other instances have been recorded in which the disturbance has been delayed until advanced age. By decades the cases in which the ages are given are as follows:

Before ten: males, none; females, 4 cases. Between ten and twenty: males, 4 cases; females, 4 cases. Between twenty and thirty: males, 10 cases; females, 13 cases. Between thirty and forty: males, 7 cases; females, 5 cases. Between forty and fifty: males, 8 cases; females, none. After fifty: males, 2 cases; females, 5 cases.

SEX. The sex of patients is about evenly divided. The totals of these records are as follows: males, 29; females, 32.

SYMPTOMS. The premonitory symptoms are of two groups, those due to pressure and those due to irritation of the environment by the epidermoidal contents. The most frequent symptom was due to encroachment upon the environment by the expanding tumor.

Pressure Symptoms. These are most frequently manifested by cough and dyspnea, less often as pain from pressure. This symptom was present in 28 cases. Cough when due to pressure is caused by irritation of the nerves. Cough of another type was caused by irritation of the bronchi when perforation was impending. When due to irritation the character of the cough is similar to that noted in pressure from aneurysm. A single case of cord paralysis has been noted.

Dyspnea, noted in 23 cases, seems to have been due to direct pressure on the trachea or bronchi, or from pressure upon and displacement of the lungs. In 3 instances death has occurred in dyspneic attacks. Dysphagia was present in 3 cases (52a, 59, 60)[1].

Irritation from the Tumor. When from causes usually unknown the tumor becomes the source of reactive inflammation, phenomena of a more violent character are induced. The cause for this irritation is not clear. The gradually increasing amount of the cyst contents probably undergoes some chemical change which inflames the sac and irritates the environment. In this they imitate the life history of wens. This similarity of reaction to that so often noted in wens was the condition that suggested the diagnosis in my patient. Pleurisy has often been diagnosed in such instances, and often exudation about the tumor has resulted which gave rise to the diagnosis of pneumonia. Often the tumor has been accidentally encountered when supposedly pleural exudates have been attacked surgically. When the bronchi are irritated, perforation into them during attacks of coughing has been noted in three cases. The attendant expectoration of other grumous material and hair has led to a positive diagnosis more often than any other factor. The invasion of the bronchus by the tumor seems to be in the nature of a pressure necrosis, often enhanced by a secondary infection of the tumor contents. The irritation of the bronchus is responded to by the production of glassy mucus. After perforation, honey-like fluid, atheromatous material, and hairs have often been observed. Clubbing of the fingers (63) and fingers and toes (57) have been noted.

Relation to Environment. The greatest possible variation in their topographical relations have been noted in dermoids of the mediastinum. The typical location of the simple dermoid is represented by my own case, a sac occupying the space between the sternum, great vessels, pericardium, and soft tissues covering the episternal notch. Every possible variation has been reported. One existed as a small egg-sized cyst in the upper part of the lung (33) near the hylus (7), while others occupied the mediastinal space and projected boldly out into the pleural cavity, and some extended from the sternum to the diaphragm, markedly displacing the lung.

[1] Figures in parentheses refer to references in the bibliography at end of article.

In one instance a retrosternal dermoid communicated by a small sinus with a similar tumor external to the sternum (39).

The relation to the surrounding structures is the factor that decides the treatment possible. For this reason these various points of attachment are worthy of note.

Connected with a bronchus, 7 cases (2, 6, 33, 36, 37, 38, 46).

Adherent to pericardium or pleura, 12 cases (7, 14, 20, 21, 26, 38, 41, 52, 63, 64, 67, 68).

Attached to lung or protruding into them, 9 cases (7, 10, 16, 19, 23, 24, 28, 45, 67).

Attached to the diaphragm. 11 cases (3, 8, 10, 22, 24, 32, 34, 41, 42, 45, 64).

Attached to the large vessels, 4 cases (1, 5, 31, 69).

Adherent to chest wall, 1 case (3).

Eleven cases extended into a lobe of a lung (6, 14, 16, 27, 33, 36, 37, 44, 47, 57, 58).

STRUCTURE. Two types may be distinguished, those in which epidermoidal tissue alone is present (28 cases) and those in which tissues from two or more of the germ layers are in evidence (25 cases).

The Simple Epidermoidal Type. These are usually a simple cyst or, at most, a conglomeration of cysts the lining of which is covered with stratified epithelium, with hair follicles, and with sweat and sebaceous glands; but a few cysts have been noted in which all appendages have been absent. Some of the simpler cysts have compartments lined with columnar epithelium, with or without cilia. The contents of cysts is usually formed by cells, fatty material, and hair. The material when in a recent state may be honey-like. A frequent striking accompaniment of this type is polypoid excrescences which project into the cavity of the cyst. These are covered with the same epidermoidal elements which line the cyst. In my case such a mass presented at once ofter the cyst was opened. Instead of or associated with such masses may be ridges of more solid tissue which traverse the cysts, dividing them partially into compartments. The mass of the projections are formed of fat.

The Teratoid Type. In the more complicated type, in addition to the epidermoidal elements, cartilage and bone are frequently found. Less often teeth have been noted. Glands, supposedly from the gut tract and from the thyroid, have been recorded. Non-striated muscle cells have been observed.

In many instances polypoid excrescences, as already noted for the simple type, project into the lumen of the tumor. These are fatty masses covered with epidermis containing fine hair, as in the simple type, but may also contain other elements. Teeth have been noted at the base of these in 6 cases (1, 32, 36, 37, 39, 42). These excrescences may form in numbers of two to six or more, and vary in size from 1 cm. square to tumors the size of a small apple.

In some of the simple epidermoid type, cuboidal or columnar epithelium is found. It is possible that these should be classified with the more complex tumors. Christian makes the suggestion that these cells may be derived from the stratum Malpighii, within which the desquamation of the other layers has occurred. The structure of these cysts recalls the structure of thymic cysts, and it is possible that this is really their relationship.

GENESIS. The origin of dermoid and teratoid tumors of the mediastinum is closely associated with the development of the thymus and thyroid glands. The close relation between the ectodermal and entodermal elements in the neck has been pointed out by Minot. That mediastinal dermoids have their origin in the upper part of the sternum is evident from their topography. Even those tumors which exhibit their greater bulk in the lower thorax retain attachments high under the sternum. In some of the reported cases, bands have extended up as far as the thyroid, suggesting an even higher origin. The existence of intestinal epithelium, as reported in one case (67), would seem greatly to complicate the problem. However, the reports are so lacking in detail that the possibility that the glandular structures observed may bear a relation to thyroid and thymus tissue (17, 20) suggests itself. Salivary gland tissue was observed in one case (68). The frequency of complicated tumors in the salivary glands is worthy of note in this connection. The association of the mixed tumors of the salivary glands and the thymic cysts would present all the elements that have been within certainty observed in mediastinal dermoids, save the teeth. The presence of teeth seems to present a barrier to a plausible explanation for the origin of the tumors under discussion, and compels us to fall back on the generalities usually employed in explaining the origin of epidermoidal or teratoid tumors in other parts of the body. Bergmann's case (39), in which a part of the tumor lay anterior to the sternum, suggests strongly that the tumor arose from a disturbance at the time of the closure of the anterior chest wall. A satisfactory explanation for the topographical relations of the more complicated type has not been presented.

DIAGNOSIS. The presence of mediastinal tumor is most often suggested by the symptoms previously noted.

Except in a few instances their size has been such as to give the usual definite physical signs of mediastinal solid bodies. In 8 instances this dulness extended lateral to the borders of the sternum, and in 9 instances it extended above the upper border of the sternum, or above either or both clavicles. In a number of instances the dulness extended over much of the side of the chest, reaching the lateral wall usually at or below the angle of the scapula. The extent and direction of the dulness above indicated presents a strongly presumptive diagnostic sign. This is made doubly so if there is a bulging above the sternum or clavicle. This had, in my case, the boggy feel of a wen, which was quite suggestive.

The examination of the sputum has given positive results in 8 cases by the discovery of hair (2, 6, 9, 27, 38, 44, 58, 66). The presence of fat or a glycerin-like fluid and squamous epithelium is strongly suggestive. The absence of pus cells may differentiate it from a simple abscess, and a negative search for tubercle bacilli is significant. In several cases aspiration has produced diagnostic evidence. Hair has been obtained by this means and presents positive evidence. Squamous keratinized cells undergoing fatty degeneration is equally positive, and fatty material, which is undergoing decomposition (13), is almost equally so. When incised the escape of honey-like material with flaky debris is strongly suggestive, as in my case. The ridges and polypoid masses covered with fine hair are readily recognizable when seen for the first time.

DIFFERENTIAL DIAGNOSIS. *Aneurysm.* Those confined to the retrosternal space or that immediately adjoining will suggest the more frequent aneurysm. The absence of beats or pulsation are the signs to be relied upon. The presence or absence of the Wassermann reaction may be of some value. Frequently the early age of the individual is of importance.

Tuberculosis. In a few instances tuberculosis has been diagnosed, owing to the dulness in the upper part of the lungs associated with expectoration. The absence of bacilli should be enough to warrant care in making such a diagnosis. Tuberculosis existed as a complication in 5 cases (6, 29, 33, 44, 57).

Empyema. The history of pain and dyspnea with the presence of fluid in the lower chest has led to error in diagnosis. Examination of the contents obtained by aspiration should be distinctive. In those rare instances in which there is a pleural exudate associated with the intrapulmonary dermoid and only the contents of the former is obtained at aspiration, error is very likely, and the operator may consider himself fortunate if he orientates himself during the course of the operation.

Malignant Tumors. The malignant tumors which are primary in the mediastinum usually run their course rapidly in contradistinction to the long history of the dermoids. However, if a dermoid becomes infected the increase in size may be even more rapid than in malignant tumors. Nevertheless, some of the dermoids have presented such urgent problems that a history was not available. Though the roentgen rays have been employed in but a few cases of mediastinal dermoids it is quite possible that the irregular masses of mediastinal malignancies might be distinguished from the more sacular dermoids. If the latter contained calcareous material, teeth or other bony structures diagnosis might be aided.

Benign Tumors. Lipomas and tumors of the thymus have presented pictures that might have been confused with mediastinal dermoids. In such cases aspiration or diagnostic incision alone could present a positive answer. With perfected technic it is to be hoped that more frequent diagnostic operations will be undertaken.

PROGNOSIS. The discussion of the prognosis is in a great measure anticipated by the natural history of the disease. Those instances in which the disease has been spontaneously fatal, hemorrhage (7.45) and dyspnea (23, 28) have been the most common causes. The size and situation, together with the reactive changes which take place in the tumor, are the determining cause. Except in very small tumors in favorable situations it is safe to say that the patient's life will be jeopardized sooner or later unless the tumor is removed or its development curtailed.

Malignant degeneration has been noted in 4 cases, carcinoma in one (69), and sarcoma in three (52b, 32, 12).

DURATION. The duration as manifested by the symptoms has varied from very brief periods to the natural life of the individual. In most instances the duration has been from one to four years. In one instance the patient died in dyspnea and no history was obtainable. In others the lesion was discovered in patients who died of some other diesase. In a number there has been a disturbance of a varied and intermittent character during the entire life of the individual.

TREATMENT. Obviously, surgical treatment alone can be of avail. Of the 27 cases operated on there was recovery in 5 (31, 39, 40, 50a, 51), improvement in 13 (13, 18, 24, 25, 29, 34, 35, 43, 48, 49, 50, 56, 57), in 4 the result was indeterminate or not stated (11, 41, 42, 63), and 5 died as the direct result of the operation (52, 53, 61, 67, 68). In most of the cases incision and drainage with the excision of the polypoid masses has been the treatment employed. In those cases reported as improved this was employed which resulted in a lessening of the size of the sac, leaving a more or less annoying sinus. A few were permanently cured by this simple means.

Total extirpation would be the ideal treatment. This was accomplished by Bastianelli (31) at a secondary operation. Total excision has been strongly recommended by von Eiselberg (52). In his 3 cases the loss of 1 by secondary hemorrhage and another by pericarditis is sufficient to show that even in the most skilled hands total removal is attended by too great a mortality to warrant the attempt at total extirpation. In many cases the relation of the tumor to surrounding structures is such that total extirpation is anatomically out of the question.

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